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**Orthopaedics
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CASE REPORT

Cervical myelopathy due to ossification of the transverse atlantal ligament: A Caucasian case report operated on and literature analysis

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KEYWORDS

Transverse atlantal ligament;
Ossification;
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Cervical myelopathy;
C1 hypoplasia

Summary One case of cervical myelopathy associated to ossification of transverse atlantal ligament (OTAL) and C1 posterior arch hypoplasia in a Caucasian adult female is reported. A 53-year-old female affected by cervical myelopathy was treated with C1 laminectomy and posterior arthrodesis. CT scan demonstrated that the distance between ossification of the ligament and anterior cortex of the posterior arch of atlas was 6,2 mm leading to consistent space reduction for spinal cord at this level. Patient underwent spinal cord decompression and fixation with C1 poliaxial screws in lateral masses and two bilateral crossing C2 laminar screws with an improvement of neurological functions at 4-years follow-up. The association between OTAL and C1 hypoplasia was reported in very few cases. The treatment with C1 laminectomy without fusion is reported in medical literature with good clinical outcome. Our patient obtained a neurological improvement at midterm follow-up with spinal cord decompression and fusion.
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Introduction

Cervical myelopathy at atlas level, in absence of trauma, is a rare pathology. In medical literature, only 14 cases of upper cervical myelopathy caused by abnormalities of

the atlas and 12 cases caused by ossification of transverse atlantal ligament (OTAL) were reported [1–20]. Only two patients out of the 26 reported, showed a rare association between atlas hypoplasia and ossification of the transverse atlantal ligament [15,20]. The incidence and aetiology of this pathology are still unclear, an important role is attributed to the ethnicity. All the described cases occurred in individuals of Asian origin [15,21]. In most cases reported clinical symptoms were characterized by severe neurological involvement due to cervical stenosis associated to OTAL [15,19,20]. In symptomatic patients the treatment is often a surgical approach in order to obtain a spinal cord decompression and recovery of neurological symptoms.

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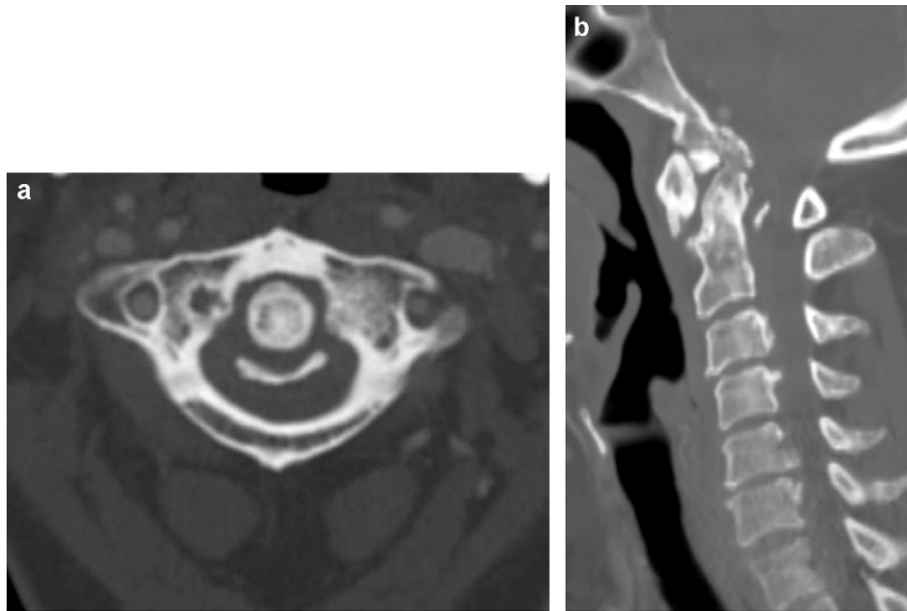


Figure 1 a, b: axial and sagittal CT scan showing the reduction of the spinal canal consequent to ossification of the transverse atlantal ligament, C1 posterior arch hypoplasia, and ossification localized between odontoid apex and occipital lamina.

Different surgical approaches to the TLA are commonly used: anterior, posterior and posterolateral [8,22,23].

We report one case of Caucasian adult female patient operated on a cervical myelopathy generated by an association of atlas hypoplasia and ossification of the transverse ligament. We discuss physiopathology and treatment options.

Clinical case

A 53-year-old Italian female presented with a 2-years history of increasing neck pain, chronic suboccipital headache and limitation of flexion and extension cervical spine range of motion. Clinically the patient presented spastic quadriparesis, worsened in the last three months, numbness of the lower and upper extremities, increased deep tendon reflexes in lower limbs, subclonic Achilles tendon reflexes, bilateral Babinski and Hoffmann signs. The patient describes progressive difficulty writing, loss of fine motor control of the hands, non-specific and diffuse weakness and abnormal sensations in the lower limbs, became very limiting in the last three months. Neurological examination revealed spastic broad-based and hesitant gait. No bowel and bladder dysfunctions were observed.

Laboratory studies showed hypertriglyceridemia: 327 mg/dl (normal range 40–170 mg/dl) and hypercholesterolemia: 234 mg/dl (normal range < 200 mg/dl). Patient presented body mass index (BMI) of 41 kg/m².

Somatosensory and motor-evoked potentials (SEPs, MEPs) were abnormal. SEPs showed functional abnormalities of the cordonal pathways rostral to C6-C7. MEPs showed alterations of the central motor conduction pathways. CT scan showed hypoplastic posterior arch of the atlas, ossification of the transverse ligament and ossification localized between odontoid apex and occipital lamina. Spinal cord compression was strictly dependent from both anterior

position of the posterior arch of C1 and ossification of the transverse ligament (Fig. 1a, b). Using a dedicated radiology medical imaging software (Kodak Direct View Picture Archiving and Communication System), we demonstrated that in the axial plane the retrodental space was 12,6 mm; the distance between ossification of the transverse ligament and anterior cortex of the posterior arch of atlas was 6,2 mm; the distance between posterior cortex of odontoid and ossification of the transverse ligament was 3.3 mm; the width of ossification in the axial plane was 3.1 mm. The major diameter of the odontoid was 11.5 mm. In the sagittal plane the height of ossification of the transverse ligament was 7.6 mm. Magnetic resonance was not performed because the patient underwent implantation of permanent pacemaker for symptomatic bradycardia ten years ago. Preoperative planning with Angio-CT demonstrated that the right vertebral artery groove at C2 level was large enough to reduce the width of the pedicle with a right dominant vertebral artery with a major diameter of 6 mm versus 2.8 mm of the left one, precluding right pedicle screw placement. The patient underwent laminectomy of C1 with posterior approach (Fig. 2) and fixation with two lateral mass polyaxial screws in the atlas, according to Harms technique [24], and two bilateral crossing C2 laminar screws according to Wright procedure [25] (SUMMIT SI OCT SYSTEM, DePuy Spine, Raynham, Massachusetts, USA). (Fig. 3a, b) C1-C2 fusion was performed with autologous iliac crest bone graft. At last follow-up (4 years) the patients showed improvement of neurological functions with persistent moderate disability and good mechanical stability of instrumentation.

Discussion

Ossification of the atlas transverse ligament and C1 posterior arch hypoplasia are rare pathologies. Only two cases of association of these two abnormalities are described in medical



Figure 2 Postoperative sagittal CT scan showing complete posterior C1 laminectomy.

literature [15,20]. Tsuruta et al. [15] in 2003 described for the first time a cervical myelopathy due to both ossification of the transverse ligament and atlas hypoplasia in one Japanese patient. Tang et al. [20] in 2010 described a similar case in an Asian patients in which was also associated an hypertrophy of the dens. An ethnic association in the development of upper cervical myelopathy due to OTAL was showed in previous studies [11,15]. All the cases described in medical literature occurred in individuals of Asian origin. In our patient the ossification of transverse ligament was associated with ossification between clivus and apex of the dens. Calcification localized into the ligamentous structure of the upper cervical spine is rare and has been reported in very few cases. Kobayashi et al. reported two cases with calcification of the alar ligament with unusual

clinical presentation [26]. Ziza et al. reported other cases with calcified deposits in the upper cervical spine [27]. Bouvet et al. described four cases with cruciform ligament ossification of the atlas [28]. Calcification of the ligaments of the cervical spine usually develops with aging and/or following minor trauma, and this disorder occurs frequently in the elderly [29]. Occipitoatlantal (C0-C1) joint is an anatomic complex structure that provides cranio-cervical range of 23–24,5° in flexion-extension [30]. We believe that because of its biomechanical characteristics the knowledge of cranio-vertebral junction is useful to recognize the specific and rare osseous abnormalities, which affect this anatomic structure. The most common congenital osseous abnormalities that involves cranio-vertebral junction are: occipitoatlantal fusion, basilar impression, occipital vertebrae, condilar apoplasia and aplasia or hypoplasia of the atlas [31]. The aetiopathogenesis of the hypoplasia of the posterior arch of the atlas is related to the embryologic development of the first cervical somites. The atlas develops from the rostral half of the first cervical sclerotome, starting from two ossification centers: the anterior and the lateral one. The development of the posterior arch of the atlas starts in the 6th week, the ossification will be complete in the 2nd or in the 3rd year of child life [15]. During the ossification process two different types of anomalies can be recognised: median clefts or lateral clefts of the posterior arch of C1, are both responsible of hypoplasia of C1 [11]. Initial size of the canal is recognised as an important factors in the development of myelopathy [32]. In a larger canal little degenerative alterations as osteophytes, ossification of ligament are tolerated without development of spinal cord compression. At the level of the atlas, as described in this case, the pathologic changes such as ossifications of the transverse ligament, can be due also to some metabolic disorders, according to some authors [9,15,20]. These disorders may include calcium-phosphate alterations, dysmetabolic syndrome, diabetes mellitus, obesity and aging. No consensus has been achieved about the role of these disorders and about their mechanism of action. Physiopathology of OTAL remains unclear. Correlation between calcification of the TAL and rheumatoid

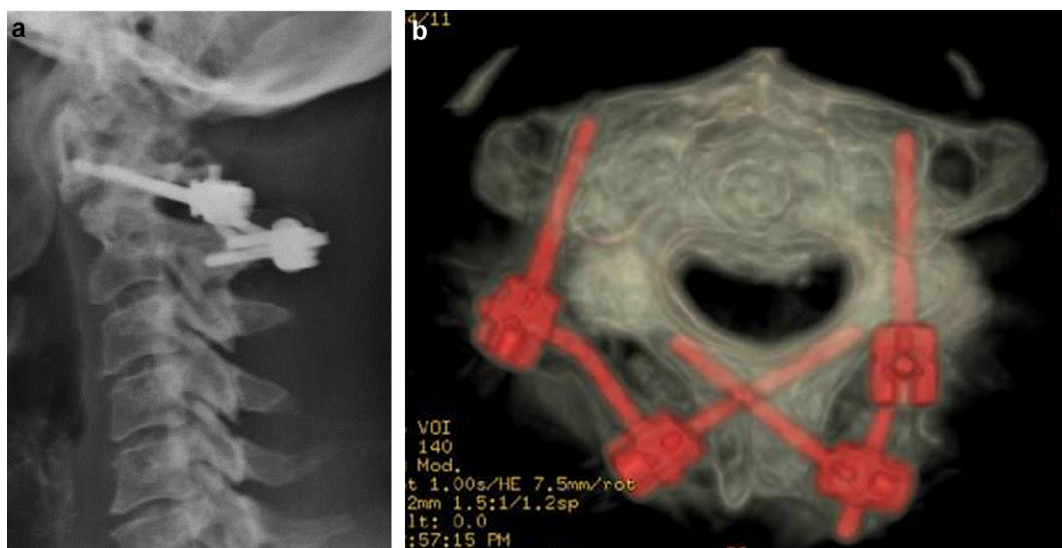


Figure 3 a, b: lateral X-Ray and 3 D CT scan showing the screws' placement in C1 lateral masses and C2 lamina.

arthritis (RA) was described by Castor et al. [33], in a study of 33 patients showing calcifications in other cervical ligaments: alar ligament, apical ligament, ligamentum flavum and posterior longitudinal ligament. However, in another study of 174 RA patients conducted by Dirheimer et al. no calcifications were mentioned [34]. Geyter et al. [35] considered the hypothesis of traumatic aetiology in combination with TAL weakness due to RA to explain the presence of ossification. Our case involves a Caucasian female adult patient, without any Asian ancestry in her lineage and no history of RA, in whom it is not possible to identify a dominant role for the ethnicity, genetic, or immunopathology related factors as responsible of the disease. As described in the medical literature a smaller posterior arch of the atlas results in a congenitally narrow spinal canal, and as a consequence restricted space available for the spinal cord, predisposing to a symptomatic spinal cord compression [11]. The normal axial diameter of the spinal canal at the level of the atlas is 21,3 mm, the retrodental space is variable from 17 to 25 mm, the normal diameter of the spinal cord is 10-12 mm, therefore secondary spinal cord compression can be suspected when the canal is less than 14 mm [36,37]. In this case the retrodental space was only 12,6 mm in the axial plane, with a distance between the odontoid and the ossified transverse ligament of 3,3 mm and as consequence space for the spinal cord of only 6,2 mm, one of the most significant described in medical literature. In fact in all cases described in the literature the spinal cord space ranges between 7 and 11 mm [15].

Different surgical approaches have been described in medical literature, considering the location of the compression. When cervical myelopathy is generated by ossification of the TLA without any other posterior abnormalities an anterior transoral approach could be useful to remove the ossified mass in front of the spinal canal [22]. Fransen et al. [23] describe posterolateral approach to obtain an anterior decompression at this level with less incidence of complications. The association between OTAL and C1 posterior arch hypoplasia could be treated with C1 posterior laminectomy to obtain adequate decompression and neurological recovery [8,15]. It is associated with a less rate of intra- and postoperative complications and a good neurological outcome. The posterior decompression could be also implemented with a C1-C2 fusion to prevent a mechanical iatrogenic instability or to treat an instability caused by the primary pathology [8,15].

In our case decompressive laminectomy of the atlas was associated with C1-C2 screw fixation, C1 poliaxial screws in the lateral masses [24], and two bilateral crossing C2 laminar screws were used [25]. C2 pedicle screw fixation was not performed because of high risk of right vertebral artery lesion due to pedicle narrowing (Fig. 4). Translaminar screws give a mechanical stability similar to pedicle fixation [38]. An accurate preoperative planning is, necessary, considering the rare pathology, the site of the lesion and finally the demanding surgical procedure. A vertebral artery injury consequent to screw placement at C1 and C2 level are described between 0 and 2,5% in medical literature with Harms technique [39]. We believe that because of the considerable variability of the vertebral artery course at the C1-C2 junction [8] the fixation of the upper cervical spine requires careful review of preoperative

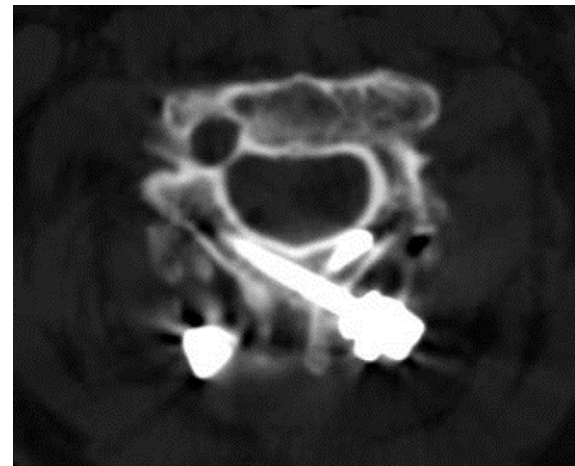


Figure 4 Axial postoperative CT scan showing the placement of the two bilateral crossing C2 laminar screws according to Wright procedure in order to preserve the right dominant vertebral artery.

Angio CT scans to optimize surgical results avoiding major complications.

Conclusion

Cervical myelopathy due to OTAL and C1 hypoplasia is an extremely rare condition. In our experience laminectomy and mechanical stability offer an opportunity for the recovery of progressive myelopathy at 4-years follow-up.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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